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A case of neglected chronic back pain in a girl with Sickle cell disease

Gowri M; Manoj K; Amudha Devi S; Kumaravel KS*; Sampathkumar D

***Corresponding Author: Kumaravel KS**
Department of Pediatrics, Govt. Mohan Kumaramangalam Medical College, Salem, India.
Email: kumaravelks10@gmail.com

Abstract

Back pain in children is due to musculoskeletal disease and trauma in the majority of the cases. The other causes of back pain in children are rarely encountered. Sickle cell disease, a blood disorder in which the sickle cells accumulate in the capillaries leading to vessel occlusion and subsequent ischemia. Here we report an eleven-year-old girl, who presented with complaints of lower backache for the past four days and difficulty in breathing for the past two days. She also had previous similar episodes of lower backache on and off for the past six years, for which she was treated as an out-patient without any specific workup or diagnosis. Musculoskeletal examination of the spine had tenderness over the lower thoracic and entire lumbar region with no erythema or swelling. High-Performance Liquid Chromatography was suggestive of Sickle Cell Disease with S-window of 66.3% and HbF of 23.6%. Magnetic Resonance Imaging (MRI) spine revealed multilevel T1/T2/STIR heterointensive lesions from T12 vertebral level to L5 level and the body height of L5 vertebra was reduced possibly due to multiple vertebral infarctions. Sickle cell disease with a vaso-occlusive crisis should also be considered as one of the differential diagnoses in children with back pain and evaluated early and appropriately to prevent long term disabilities resulting from complications like vertebral collapse.

Keywords

Vaso-occlusive crisis; Infarction; Sequestration; Collapse of vertebra.

Introduction

Low back pain is a common problem in adults. Back pain in children is due to musculoskeletal disease and trauma in the majority of the cases. The other causes of back pain in children will include infection, neoplastic lesions, rheumatologic diseases, and miscellaneous causes like a vaso-occlusive crisis, nephrolithiasis, and osteomyelitis [1]. Even though most cases of back pain have benign etiology, it is important to evaluate these children for any underlying serious pathology.

Sickle cell disease (SCD) is a blood disorder that follows an autosomal recessive pattern of inheritance. SCD is caused by a point mutation in the sixth position of a beta-globin chain of hemoglobin, where hydrophilic glutamic acid is replaced by hydrophobic amino acid valine. The beta-globin gene is found on chromosome 11. SCD results when both beta-globin chains are abnormal-either homozygous (HbSS) or heterozygous (combined with other abnormal hemoglobins, such as hemoglobin C [HbSC] or beta thalassemia [HbS-thal]) [2]. The most common form of SCD is sickle cell anemia or HbSS disease. The combination of a sickle beta-globin chain with a normal one (HbSA), results in the sickle cell trait.

The abnormal hemoglobin morphology reduces the flexibility of Red Blood Cells (RBC) so that they become rigid, sickle-shaped in the setting of low oxygen tension. Physiological stress, high altitude, temperature changes and dehydration are the most common conditions that can precipitate sickling of red blood cells in patients with SCD [3]. These sickled cells are not able to transform back to their normal biconcave shape, even though oxygen tension is normal. The sickle cells accumulate in the capillaries leading to vessel occlusion and subsequent ischemia. SCD is more common in people from the Middle East and the eastern Mediterranean region, but most prevalent in those of African origin. In this case report, we present a girl with backaches as the only manifestation of SCD.

Case Histroy

An eleven-year-old girl, the third born of third-degree consanguineous parents presented with complaints of lower backache for the past four days and difficulty in breathing for the past two days. The child also had a complaint of difficulty in walking due to lower backache. The child had previous similar episodes of lower backache on and off for the past six years, for which she was treated as an out-patient without any specific workup or diagnosis. The child also had no history of previous hospitalization or blood transfusion. Antenatal and natal histories were uneventful. The child was developmentally normal for age. She was immunized up to age. Despite parental consanguinity, there were no similar complaints, and no blood transfusion history was present in other family members. There was no contact history with tuberculosis.

On general examination, the child was found to be in pain, conscious and oriented, had signs of marked pallor and jaundice. The height and the weight of the child were below the 3rd percentile for age. The child was afebrile and hemodynamically stable. The child was tachypneic and mild sub costal retractions were present. Abdominal examination revealed splenomegaly with spleen palpable 5 cms below the left costal margin. Cardiovascular and neurological examinations were normal. Musculoskeletal examination of the spine had tenderness over the lower thoracic and entire lumbar region with no erythema or swelling. She had limited movements of the trunk, with pain during forward and lateral flexion.

On investigations, Blood Counts showed a Total Count of 11,000 cells/mm^3 (Polymorphs -69%, Lymphocytes -30%, Eosinophils -1%), Hb% was 6.4 gms%, Packed Cell Volume was 18%, total bilirubin was 2.4 gms%, direct bilirubin was 0.9 mgs%, and lactate dehydrogenase (LDH) was 892 mg%. Direct Coomb's test was negative. Reticulocyte count was 1.5%. The peripheral smear (Figure 1) showed severe microcytic, hypochromic anemia, anisiopoikilocytosis, few elliptical and targeted cells, and occasional sickle-like cells. The peripheral smear picture was suggestive of hemolytic anemia. A sickling test with 2%

sodium meta-bisulphite demonstrated plenty of sickled RBCs (Figure 2).

High Performance Liquid Chromatography (HPLC) was suggestive of SCD with S-window of 66.3% and HbF of 23.6%. HPLC analysis of the parents revealed that the father and the mother of the child were having sickle cell traits. Ultrasonographic examination of the abdomen showed moderate splenomegaly. Magnetic Resonance Imaging (MRI) spine revealed multilevel T1/T2/STIR heterointensive lesions from T12 vertebral level to L5 level and the body height of L5 vertebra was reduced possibly due to multiple vertebral infarctions (Figure 3).

With blood transfusion and other supportive measures, the girl improved. Narcotic analgesics were given for acute pain relief. The parents were counseled regarding the disease. The girl was discharged after 2 weeks in a stable condition and was advised to take ibuprofen for pain whenever necessary.

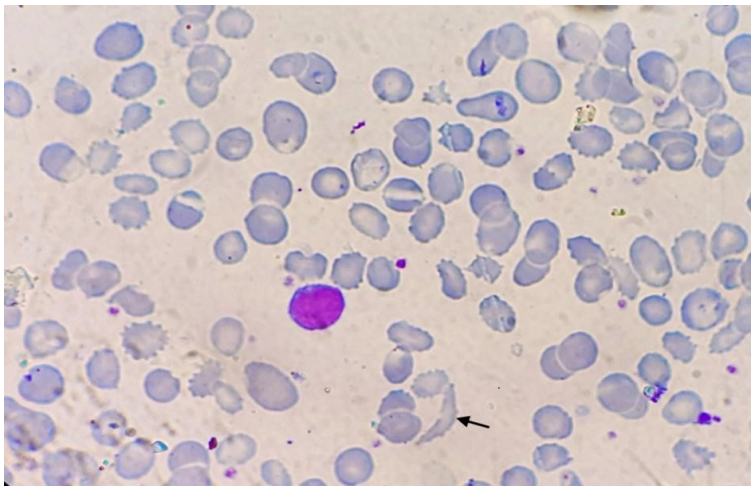

Figure 1: Peripheral smear study showing microcytic hypochromic RBCs, few elliptical cells, few target cells, and occasional sickle cells (arrow).

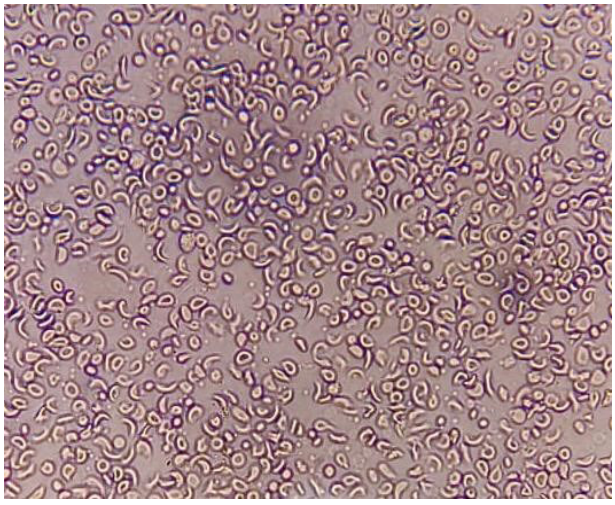

Figure 2: Sickling test with sodium meta-bisulphite demonstrating plenty of sickled RBCs.

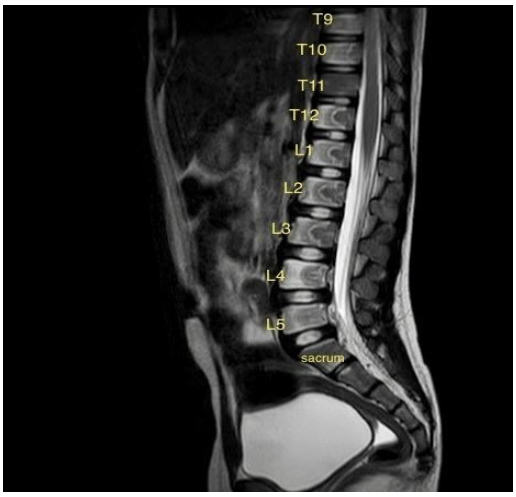

Figure 3: T2 weighted MRI image of the lateral spine revealed multilevel heterointensive lesions and height reduction of L5 vertebra.

Discussion

Children with SCD can present with painful vaso-occlusive crisis, osteomyelitis, stress fractures, orbital compression syndrome, dental problems, vertebral collapse and bone marrow necrosis [3]. The acute painful vaso-occlusive crisis is the most common acute clinical presentation of SCD in children. It is due to sickled red blood cells causing microvasculature occlusion resulting in chronic tissue ischemia and infarction. It can occur in any bone with active marrow most commonly in the spine and knee. Among the spine affected cases, two-thirds have lumbosacral involvement, while an additional 20% have thoracic spine involvement. Chronic bone problems in SCD include avascular necrosis, chronic arthritis, osteopenia and osteoporosis.

Radioisotope bone scans can reliably recognize acute phase areas of infarction. A bone marrow scan or MRI is more sensitive in detecting infarcts [4]. The reported radiological signs of bone infarction include the "fish mouth vertebra" sign, "vanishing vertebra", coarse trabecular pattern, persistent anterior vertebral notching, and biconcavity of the bodies, step ladder effect, the massive collapse of the "central" and compression deformities. There can be multiple osteolytic bone lesions involving the lumbar vertebra with a decrease in the height of the affected vertebral bodies as presented in this case.

The treatment of vertebral infarcts and destruction in SCD patients is chiefly conservative because of the high incidence of intra and peri-operative complications, including acute respiratory syndrome, vaso-occlusive crisis and increased risk of transplant failure [3]. There is an increased risk of post-operative wound infection due to impaired immune function and splenic dysfunction. Conservative management includes rest and symptomatic treatment of acute painful episodes. Pain medications and muscle relaxants can be useful but chronic use is associated with side effects. Thoracic or lumbar Orthosis can be used instead of such chronic medication [5]. Lumbar jackets also improve patient ability to ambulate and improve their ability to be given outpatient management. Surgical treatment can be done when spinal pathology secondary to SCD occurs in a young patient with otherwise well-controlled SCD (6). Surgery should be considered if the children develop kyphotic deformity.

Conclusion

Sickle cell disease with vaso-occlusive crisis should also be considered as one of the differential diagnoses in children with back pain and evaluated early and appropriately to prevent long term disabilities resulting from complications like vertebral collapse.

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Authors Information: Gowri M; Manoj K; Amudha Devi S; Kumaravel KS*; Sampathkumar D
Department of Pediatrics, Govt. Mohan Kumaramangalam Medical College, Salem, India.

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